MELANOMA AND MELANOCYTIC NAEVI

PAINFUL PAPULO-NODULES WITH ZONIFORM DISPOSITION REVEALING REED'S SYNDROME

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Background: Cutaneous and familial uterine leiomyomatosis or Reed syndrome is a rare hereditary disease, with autosomal dominant inheritance, defined by the presence of multiple cutaneous and uterine leiomyoma, and predisposition to the development of renal cell carcinoma. It is secondary to a mutation in the gene encoding fumarate hydratase. We report a case discovered after a 23-year diagnostic wandering, and particular by the zosteriform arrangement of cutaneous lesions.

Observation: A 50-year-old patient who has been suffering from papulo-nodules of the right cheek for 23 years, gradually increasing in number, and having become painful to touch and exposed to cold since 3 years. Clinical examination found numerous firm erythematous papulonodules (>10) grouped in zosteriform arrangement, painful to the touch. The cutaneous biopsy showed a proliferation of smooth muscle cells, in favor of cutaneous leiomyomatosis. By carefully questioning with the patient, she reveals that her and her sisters underwent hysterectomy for uterine leiomyoma. Her son had similar skin lesions, and one of her sisters died of kidney cancer. The diagnosis of Reed syndrome was then established. Renal MRI to screen for cancer was normal. An analgesic treatment with gabapentin has been instituted. A renal MRI screening is planned annually.

Key message: Reed syndrome is a rare genodermatosis with risk of renal cancer. The role of the dermatologist comes for the early diagnosis of this entity and screening for renal cancer.